Fibrous Dysplasia of the Proximal Part of the Femur

LONG-TERM RESULTS OF CURETTAGE AND BONE-GRAFTING
AND MECHANICAL REALIGNMENT*

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ABSTRACT: We reviewed the long-term outcomes of treatment of fibrous dysplasia of the proximal part of the femur in twenty-two patients (twenty-seven femora). There were fifteen male patients and seven female patients. Patients who had monostotic disease had no involvement of the calcar femorale, fewer microfractures, less deformity, and stronger bone that could support internal fixation. Patients who had polyostotic disease had frequent involvement of the calcar femorale; more microfractures; severe deformity, including shepherd's crook deformity; and, in many instances, bone that could not support internal fixation. Twenty-two of the twenty-seven femora had a microfracture at the time of the initial presentation.

At least one osteotomy was performed in four femora that had monostotic disease and in nine femora that had polyostotic disease. Curettage and cancellous or cortical bone-grafting did not appear to have any advantage compared with osteotomy alone in the treatment of symptomatic lesions, as all grafts resorbed with persistence of the lesion. At the time of the latest follow-up evaluation, no lesion had been eradicated or had decreased in size.

A satisfactory clinical result was achieved in twenty patients (twenty-four femora): nine who had monostotic disease and eleven who had polyostotic disease. Two patients who had polyostotic disease and an endocrinopathy (one of whom had bilateral involvement) had an unsatisfactory result. All three femora in these two patients had a neck-shaft angle of less than 90 degrees at the time of the most recent follow-up evaluation.

Varus deformity of the proximal part of the femur is best treated with valgus osteotomy and internal fixation early in the course of the disease. If the calcar of the femoral neck is involved or if the quality of the bone is such that internal fixation is not possible, a medial displacement valgus osteotomy can provide a more mechanically favorable position for healing of the microfracture.

Fibrous dysplasia is an anomaly characterized by widening of the affected bone with cortical thinning and by the presence of fibro-osseous tissue in the interior of the bone. There also may be areas with islands of cartilage or cysts, and some lesions may be expansile in nature. Fibrous dysplasia of bone is an enigma with no known cure. The progression of the lesion is erratic; strong mechanical forces acting on the weakened proximal part of the femur result in a progressive varus and bowing deformity in most patients[4,9]. Treatment currently consists of curettage and bone-grafting in an attempt to eradicate the lesion as well as valgus realignment of the proximal part of the femur to correct the progressive varus deformity, which may range from mild coxa vara to a marked shepherd's crook deformity[4,6,9,10,17,19-21]. The degree of the deformity depends on the extent and location of the lesion and on whether the lesion is monostotic or polyostotic.

Harris et al., in their classic review of the natural history of fibrous dysplasia, reported involvement of the femur in 92 per cent of thirty-seven patients who had a polyostotic lesion. However, the orthopaedic literature contains only a few reports that deal with the treatment of fibrodysplastic lesions of the proximal part of the femur and even fewer with long-term follow-up. The purpose of the present study was to determine the effectiveness of curettage and bone-grafting in eradicating these lesions and to evaluate the long-term effectiveness of mechanical realignment of the proximal part of the femur.

Materials and Methods

We reviewed the case records and roentgenograms of all patients who were seen at the Alfred I. duPont Institute between 1940 and 1992 with a biopsy-proved diagnosis of fibrous dysplasia about the hip. Only patients who had involvement of the proximal part of the femur and who had been followed for a minimum of two years were included in the study; twenty-two patients (twenty-seven femora) met these criteria.

The operative notes were reviewed with regard to the

indications, techniques, and results. The location and appearance of the fibrodysplastic lesion at the time of both the initial evaluation and the most recent follow-up visit were recorded. Because the extent of the lesion tends to change with growth of the patient, any change in size was noted. At the time of the latest follow-up examination, nine femora had thinning of the cortex and a change in the size of the lesion, but these changes were difficult to quantitate as some were secondary to growth as opposed to actual progression of the lesion. A shepherd's crook deformity — defined as increased bowing of the proximal part of the femur with a varus deformity of the femoral neck (a neck-shaft angle of less than 90 degrees) — was noted in six femora; all six deformities were associated with a lesion that involved the entire proximal part of the femur or the neck and the intertrochanteric region. All complete fractures and microfractures were recorded. As the fibrodysplastic lesions themselves are not inherently painful, a microfracture was assumed to have occurred when there was a clinical history of a limp and an acute onset of sharp pain in the region of the hip, with or without a thin radiolucent line in the bone as seen on roentgenograms. The neck-shaft angle was measured on anteroposterior roentgenograms made at the time of the initial visit or before the osteotomy and at the time of the most recent follow-up.

The clinical results initially were evaluated with use of the Iowa hip score, but we found that this system was not applicable to our patient population. We therefore used a minimum of two clinical criteria to categorize the results as satisfactory or unsatisfactory. These criteria included pain (no pain or mild pain with activity was considered satisfactory and more severe pain, unsatisfactory), range of motion (a functional range was considered satisfactory and restricted motion, unsatisfactory), ability to perform activities of daily living (unlimited ability was considered satisfactory and limited ability, unsatisfactory), antalgic limp (no limp or a mild limp with activity was considered satisfactory and a worse limp, unsatisfactory), and overall ability to function in society (unlimited ability was considered satisfactory and limited ability, unsatisfactory).

In general, the indications for the various operative procedures were consistent. Curettage and bone-grafting was performed in an attempt to eradicate the lesion, often in conjunction with a biopsy. A valgus osteotomy was performed when the proximal part of the femur had a progressive varus deformity of 10 degrees or more or when it was thought that valgus positioning would provide better mechanical alignment for healing of a fracture. The choice of internal fixation varied over the fifty-two-year period of the study and often depended on the quality of the bone and its ability to support internal fixation. Pins, plates, wires, screws, and rods were used accordingly. When the lesion involved the calcar or when the quality of the bone prohibited the use of internal fixation, a medial displacement val-

gus osteotomy, as described by McMurray, was performed. Postoperatively, a one and one-half hip-spica cast was used for all patients.

The patients were divided into two groups. Those in Group I had monostotic involvement (a single known lesion in the proximal part of the femur) and those in Group II had polyostotic involvement (at least one other bone, in addition to the femur, was affected). Five patients in Group II had involvement of both femora; none of these patients needed operative treatment of both hips. The femora were subdivided further according to the initial location of the lesion. Lesions involving the entire proximal part of the femur were categorized as Subgroup A; those involving the femoral neck only, as Subgroup B; those involving both the femoral neck and the intertrochanteric region, as Subgroup C; and those involving the intertrochanteric region only, as Subgroup D.

At the time of the latest follow-up, the lesion had enlarged in six femora in Group I and in three in Group II.

Group I: Monostotic Disease

There were nine patients (nine femora) in this group (Table I). Seven patients were male and two were female. The right femur was involved in six patients and the left femur was involved in three. The average age at the time of the diagnosis of the fibrous dysplasia was 8.6 years (range, 4.3 to 14.2 years). The average age at the time of the first visit to our institution was 11.1 years (range, 4.3 to 18.0 years).

The presenting symptoms for all nine patients were pain and an antalgic limp, which are indicative of a microfracture according to our definition; in four patients, the microfracture was seen on roentgenograms. Two patients had involvement of the entire proximal part of the femur (Subgroup A), one had involvement of the femoral neck only (Subgroup B), three had involvement of both the femoral neck and the intertrochanteric region (Subgroup C), and three had involvement of the intertrochanteric region only (Subgroup D). One of the patients in Subgroup A had a shepherd's crook deformity.

The average age at the time of the most recent follow-up examination was 22.2 years (range, 9.0 to 33.9 years), and the average duration of follow-up was 13.6 years (range, 2.5 to 28.5 years).

Group II: Polyostotic Disease

There were thirteen patients (eighteen femora) in this group. Five patients (Cases 12 and 13, 14 and 15, 17 and 23, 18 and 19, and 21 and 22) had the lesion bilaterally. Eight patients were male and five were female. The average age at the time of the diagnosis was 7.8 years (range, 1.5 to 13.9 years). The average age at the time of the first visit to our institution was 11.1 years (range, 3.5 to 26.1 years).

Twelve patients were first seen because of pain and an antalgic limp. One patient was seen because of a wad-

TABLE I
DATA ON THE TWENTY-TWO PATIENTS (TWENTY-SEVEN FEMORA)

Case	Indication/Treatment/Age (yrs.)				Durat. of/ Age at Latest Follow-up (yrs.)	Neck-Shaft Angle* (degrees)	Size of Lesion at Latest Follow-up	Result	Comments
	Initial	2nd	3rd	≥4th					
Monostotic disease									
Group IA: entire prox. part of femur									
1	Microfract./cast/4.3	Persist. lesion/curet. & bone graft/7.0	—	—	4.7/9.0	146/139	Larger	Satis.	—
2	Microfract./cast/4.5	Microfract./cast/12.0	Microfract./cast/18.0	Shepherd's crook deform./curet. & bone graft & valgus osteot. with hip-screw/29.0	28.5/33.0	90/130	Larger	Satis.	Microfracts. healed in varus; shepherd's crook deform.
Group IB: femoral neck									
3	Microfract./curet. & bone graft/14.2	Microfract./non-weight-bearing/15.2	Microfract./fibular graft/22.2		19.7/33.9	134/134	Larger	Satis.	Graft resorbed
Group IC: femoral neck & intertrochanteric region									
4	Microfract./cast/6.0	Intertrochanteric fract./cast/13.0	Varus deform./valgus osteot. with Zickel nail/13.6	—	25.2/31.2	70/120	Same	Satis.	—
5	Microfract./curet. & bone graft/4.4	Persist. lesion/curet. & bone graft/7.5	—	—	4.6/9.0	174/142	Larger	Satis.	—
6	Microfract./curet. & bone graft/13.5	Persist. lesion/curet. & bone graft/15.8	—	—	2.5/16.0	144/144	Larger	Satis.	—
Group ID: intertrochanteric region									
7	Microfract./cast/11.4	Microfract./cast/13.4	Varus deform./valgus osteot. with hip-screw/15.7	—	9.3/20.7	115/147	Same	Satis.	Microfracts. healed in varus
8	Microfract./curet. & bone graft/6.0	Intertrochanteric fract./cast/10.0	Persist. lesion/curet. & bone graft/11.1	Microfract./cast/11.9; varus deform./valgus osteot. with blade-plate/13.4; microfract./valgus osteot. with hip-screw/18.5	22.9/28.9	110/142	Larger	Satis.	Blade-plate did not extend into femoral head
9	Microfract./curet. & bone graft/13.5	—	—	—	4.8/18.3	135/135	Same	Satis.	—
Polyostotic disease									
Group IIA: entire prox. part of femur									
10	Microfract./cast/4.2	Persist. lesion/curet. & bone graft/5.5	Shepherd's crook deform./med. displace. valgus osteot./9.0	—	8.8/13.0	75/50	Larger	Unsatis.	Loss of correct.
11	Varus deform., microfract./valgus osteot./7.0	Varus deform./med. displace. valgus osteot./13.1	—	—	17.6/24.6	95/95	Larger	Satis.	Expansile lesion
12	Microfract./valgus osteot./9.0	Microfract./valgus osteot./14.0	Shepherd's crook deform./valgus osteot. with Küntscher nail/23.0	Microfract./cast/26.1	31.8/40.8	NA/95	Same	Satis.	Acromegaly; bilat. with Case 13
13	Lesion seen on roentgenogram	—	—	—	31.8/40.8	NA/95	Same	Satis.	Acromegaly; bilat. with Case 12

TABLE I *(Continued)*
DATA ON THE TWENTY-TWO PATIENTS (TWENTY-SEVEN FEMORA)

Case	Indication/Treatment/Age *(yrs.)*				Durat. of/ Age at Latest Follow-up *(yrs.)*	Neck-Shaft Angle* *(degrees)*	Size of Lesion at Latest Follow-up	Result	Comments
	Initial	2nd	3rd	≥4th					
14	Waddling gait, varus deform./med. displace. valgus osteot./9.7	Microfract./cast/10.3	Microfract./cast/14.7	Varus deform./med. displace. valgus osteot./15.1; microfract./cast/15.9; micro-fract./cast/18.7	15.0/24.7	60/66	Same	Unsatis.	McCune-Albright synd.; bilat. with Case 15
15	Lesion seen on roent-genogram	—	—	—	15.0/24.7	110/64	Same	Unsatis.	McCune-Albright synd.; bilat. with Case 14
16	Microfract./curet. & bone graft/8.5	—	—	—	19.8/28.3	136/136	Same	Satis.	Acromegaly
Group IIB: femoral neck									
17	Lesion seen on roent-genogram	—	—	—	9.5/20.4	148/120	Same	Satis.	McCune-Albright synd.; bilat. with Case 23
Group IIC: femoral neck & intertrochan-teric region									
18	Microfract./cast/12.7	—	—	—	8.1/20.8	119/114	Same	Satis.	Bilat. with Case 19
19	Microfract./cast/13.3	Shepherd's crook de-form./med. displace. valgus osteot./13.6	—	—	8.1/20.8	120/137	Same	Satis.	Bilat. with Case 18
20	Microfract./cast/9.7	Intertrochan-teric fract./cast/14.3	Microfract./cast/14.7	Shepherd's crook deform./valgus osteot./15.7	7.9/17.6	125/135	Same	Satis.	—
21	Microfract./cast/2.0	Microfract./curet. & bone graft/4.8	Varus deform./med. displace. valgus osteot./7.0	Persist. lesion/curet. & bone graft/8.0; varus deform./valgus osteot. & curet. & bone graft/9.7; varus de-form./valgus osteot./16.0	41.3/43.3	100/136	Same	Satis.	Bilat. with Case 22
22	Lesion seen on roent-genogram; microfract./cast/11.1	—	—	—	41.3/43.3	115/115	Same	Satis.	Bilat. with Case 21
23	Microfract./cast/7.2	Persist. lesion/curet. & bone graft/8.5	Persist. lesion/curet. & bone graft/9.7	Microfract. & varus deform./med. displace. valgus osteot./11.9	13.2/20.4	86/118	Same	Satis.	McCune-Albright synd.; bilat. with Case 17
24	Microfract./cast/5.4	Varus deform./curet. & bone graft & valgus osteot. with blade-plate/10.8	Microfract./curet. & bone graft & valgus osteot. with hip-screw/16.2	Microfract./valgus osteot. & curet. & bone graft with hip-screw/17.2; persist. lesion/curet. & bone graft/19.5	17.3/22.7	135/140	Same	Satis.	Hip-screw did not extend into fem-oral head
25	Microfract./cast/1.5	Microfract./cast/2.5	Persist. lesion/curet. & bone graft/4.6	Persist. lesion/curet. & bone graft/6.3	6.4/7.9	155/115	Larger	Satis.	McCune-Albright synd., shepherd's crook deform., microfracts. healed in varus
26	Microfract./non-weight-bearing/13.9	—	—	—	2.0/15.9	136/136	Same	Satis.	—
27	Microfract./cast/11.1	—	—	—	11.2/22.3	115/115	Same	Satis.	McCune-Albright synd.

*At the initial visit or before the osteotomy/at the latest follow-up evaluation. NA = not available.

FIG. 1-A   FIG. 1-B   FIG. 1-C

Figs. 1-A through 1-E: Case 8. Serial roentgenograms of a patient who had monostotic disease.
Fig. 1-A: When the patient was 6.0 years old, a lesion was noted in the intertrochanteric area.
Fig. 1-B: After two curettage and bone-grafting operations, there was extension of the lesion into the femoral neck and the subtrochanteric region.
Fig. 1-C: Following a microfracture, a varus deformity developed. This was treated with a valgus osteotomy and blade-plate fixation. The blade-plate did not traverse the entire length of the femoral neck.

dling gait, and a microfracture was also found. This patient had bilateral involvement and severe coxa vara. Eleven femora had a microfracture that was seen on the roentgenograms. One patient who had bilateral involvement had no symptoms referable to the right hip; the lesion was identified in the right femur when roentgenograms of the pelvis were made to evaluate the symptomatic, left hip, which had a microfracture. Seven femora had involvement of the entire proximal part (Subgroup A); one, of the neck only (Subgroup B); and ten, of both the neck and the intertrochanteric region (Subgroup C). A shepherd's crook deformity was seen in five femora (two in Subgroup A and three in Subgroup C).

The average age at the time of the latest follow-up examination was 23.3 years (range, 7.9 to 43.3 years), and the average duration of follow-up was 15.0 years (range, 2.0 to 41.3 years).

Results

Group IA: Monostotic Disease with Involvement of the Entire Proximal Part of the Femur

Both patients (one male and one female) in this group were first seen because of pain and a limp caused by a microfracture (Table I). Both of these patients were managed with a one and one-half hip-spica cast,

and both had healing without deformity.

The male patient (Case 1) had persistence of the lesion and was managed with curettage and bone-grafting. At the most recent follow-up evaluation, the neck-shaft angle measured 139 degrees compared with the initial angle of 146 degrees.

The female patient (Case 2) had two subsequent microfractures; both were treated with a one and one-half hip-spica cast, and both healed in a varus position. When she was 29.0 years old, she was seen with a neck-shaft angle of 90 degrees and a shepherd's crook deformity. She was managed with curettage and bone-grafting followed by a valgus osteotomy with a sliding hip-screw. At the time of the latest follow-up, when she was 33.0 years old, the neck-shaft angle was 130 degrees.

At the time of the latest follow-up, the lesion was larger in both patients. Neither had pain in the hip or needed aids for walking. Both patients had a satisfactory result.

Group IB: Monostotic Disease with Involvement of the Femoral Neck

One male patient was initially seen because of pain and a limp when he was 14.2 years old, at which time

a lesion was noted on the roentgenogram (Table I). Although he was presumed to have a microfracture because of the symptoms, it could not be identified on the roentgenogram and the calcar of the femoral neck was intact. He was managed with curettage and bone-grafting. One year later, he sustained another microfracture, but deformity of the femoral neck did not develop. He was managed with non-weight-bearing with use of crutches. Seven years later, the patient had another microfracture. This was treated with an autologous fibular strut graft, as described by Enneking[5].

At the time of the most recent follow-up, when the patient was 33.9 years old, the neck-shaft angle remained unchanged at 134 degrees, but the lesion persisted and was slightly larger. The patient had no pain in the hip and walked without aids. He had a satisfactory result.

Group IC: Monostotic Disease with Involvement of the Femoral Neck and the Intertrochanteric Region

Three male patients were in this group (Table I). The first patient (Case 4) initially was seen because of a microfracture, which was treated with a one and one-half hip-spica cast. He later sustained a displaced intertrochanteric fracture, due to trauma; the fracture was reduced and was treated with a one and one-half hip-spica cast. Six months later, the neck-shaft angle was 70 degrees; a valgus osteotomy was performed with use of a Zickel nail. At the time of the latest follow-up evaluation, the neck-shaft angle was 120 degrees.

The two remaining patients initially were seen because of pain and an antalgic limp. Both had a presumed microfracture, which was treated with curettage and bone-grafting. The lesion persisted in both patients, necessitating repeat curettage and bone-grafting. At the time of the most recent follow-up, one patient (Case 5) had a neck-shaft angle of 142 degrees compared with an initial angle of 174 degrees. The other patient (Case 6) had a neck-shaft angle of 144 degrees, which was the same as the initial measurement.

At the time of the latest follow-up, the lesion persisted in all three femora and had increased in the two that had been treated with curettage and bone-grafting. None of the three patients had problems with walking, and all were satisfied with the result.

Group ID: Monostotic Disease with Involvement of the Intertrochanteric Region

This group comprised two male patients and one female patient, all of whom initially were seen because of a microfracture (Table I). The first patient (Case 7) was managed with a one and one-half hip-spica cast twice, but a varus deformity of the proximal part of the femur subsequently necessitated a valgus osteotomy with internal fixation. The second patient (Case 8) had two curettage and bone-grafting procedures and a subsequent valgus osteotomy with use of a blade-plate (Figs. 1-A through 1-E). The blade-plate did not traverse the entire length of the femoral neck and a progressive varus deformity developed, necessitating a repeat valgus osteotomy and fixation with a hip-screw that traversed the entire femoral neck and entered the femoral
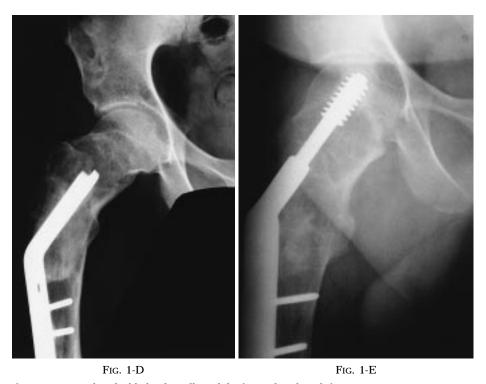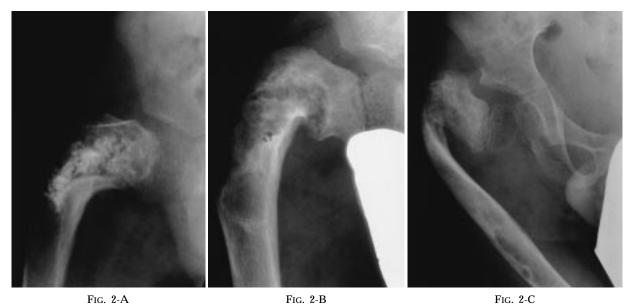

FIG. 1-D   FIG. 1-E

Fig. 1-D: A microfracture proximal to the blade-plate allowed the femoral neck to deform into varus.
Fig. 1-E: A second valgus osteotomy was performed and was stabilized with a compression hip-screw extending into the femoral head.

<div align="center">Fig. 2-A   Fig. 2-B   Fig. 2-C</div>

Figs. 2-A through 2-E: Case 10. Serial roentgenograms of a patient who had polyostotic disease and endocrinopathy.
Fig. 2-A: When the patient was 5.5 years old, curettage and bone-grafting was performed.
Fig. 2-B: The lesion persisted one year later, and the beginning of a shepherd's crook deformity was noted.
Fig. 2-C: When the patient was 7.6 years old, a marked shepherd's crook deformity was noted.

head. The third patient (Case 9) was managed with curettage and bone-grafting.

At the time of the latest follow-up, one lesion was larger and the other two were approximately the same size. The average neck-shaft angle was 141 degrees (135, 142, and 147 degrees) compared with the initial angle of 120 degrees (110, 115, and 135 degrees). All three patients walked without aids, and all had a satisfactory result.

Group IIA: Polyostotic Disease with Involvement of the Entire Proximal Part of the Femur

There were five patients (four male and one female), with seven involved femora, in this group (Table I). Three patients had an endocrine abnormality. Two patients (Cases 12 and 13 and Cases 14 and 15) also had involvement of the proximal part of the contralateral femur, but this was not treated. All five patients had a microfracture (in five femora). All of the fractures healed, but a progressive varus deformity developed in four. In two femora (Cases 10 and 12), this progressed to a shepherd's crook deformity (Figs. 2-A through 2-E). These four femora were treated with a valgus osteotomy, with medial displacement in three. Three femora needed more than one osteotomy; internal fixation of the site of the osteotomy was not technically feasible in any of them. Two patients had curettage and bone-grafting.

At the time of the most recent follow-up, two lesions were larger and five were approximately the same size. The average neck-shaft angle was 86 degrees (range, 50 to 136 degrees) compared with the initial angle of 95 degrees (range, 60 to 136 degrees). Two patients were able to walk with crutches, and one used a cane. One patient (Cases 14 and 15), who had McCune-Albright syndrome, had such extensive involvement of the upper and lower extremities that she could not walk, and she used a wheelchair. Another patient needed no aids for walking. Three patients had a satisfactory result and two had an unsatisfactory result.

Group IIB: Polyostotic Disease with Involvement of the Femoral Neck

The one female patient in this group had McCune-Albright syndrome and bilateral involvement (Table I). The right femur (Case 17) never fractured or needed treatment. The initial neck-shaft angle was 148 degrees. At the time of the latest follow-up evaluation, when the patient was 20.4 years old, the neck-shaft angle was 120 degrees and the lesion persisted although it was no larger. The hip was not painful, and the patient did not need aids for walking. The left femur (Case 23) was classified as Group IIC. The patient had a satisfactory result.

Group IIC: Polyostotic Disease with Involvement of the Femoral Neck and the Intertrochanteric Region

This group comprised eight patients (four male and four female), three of whom had bilateral involvement (Table I). Two patients (Cases 18 and 19 and Cases 21 and 22) had Group-IIC involvement of both femora, but only one hip in each patient was treated operatively. Both of these hips (Cases 19 and 21) had a medial displacement valgus osteotomy because of the varus deformity, which had progressed to a shepherd's crook deformity in one (Case 19). The third patient had Group-IIB involvement of one femur (Case 17), which was not treated, and Group-IIC involvement of the other femur (Case 23), which was treated with curettage and bone-grafting

Fig. 2-D: Roentgenogram made after a medial displacement valgus osteotomy.
Fig. 2-E: One year after the osteotomy, there is a solid union. The patient was temporarily lost to follow-up but returned at the age of 13.0 years because the shepherd's crook deformity had recurred.

twice and a medial displacement valgus osteotomy.

All ten femora in this group had a microfracture. In one femur (Case 26), the symptoms related to the microfracture resolved following a trial period of non-weight-bearing. Three femora (Cases 18, 22, and 27), each of which had one microfracture, were treated with a one and one-half hip-spica cast. All three femora had a mild varus deformity, but it was not progressive and none had any additional treatment.

Four femora were treated with curettage and bone-grafting; three also had a valgus osteotomy, which included medial displacement in two. Of these femora, one (Case 21) had five operations because of persistent microfractures and a progressive varus deformity. Internal fixation was used in one femur, but it needed to be revised as the first hip-screw did not extend the length of the femoral neck. The lesion did not heal in any of these four patients.

Two femora (Cases 19 and 20) that had a shepherd's crook deformity were treated with a valgus osteotomy, which included medial displacement in one (Case 19). Another femur (Case 25) had repeated microfractures that healed in varus; curettage and bone-grafting were performed twice because of persistence of the lesion, but this exacerbated the varus deformity. A shepherd's crook deformity was noted at the time of the most recent follow-up.

At the time of the latest follow-up, the average neck-shaft angle was 126 degrees (range, 114 to 140 degrees) compared with the initial angle of 121 degrees (range, 86 to 155 degrees). The lesion remained the same size in nine femora and increased in one. All patients were able to walk without aids, and all had a satisfactory result.

Growth Arrest

Four femora (Cases 2, 6, 17, and 27) had what appeared to be a disturbance in the growth of the lateral aspect of the capital femoral physis, resulting in apparent elevation of the medial side of the capital epiphysis with a lateral tilt. In two of these femora, the fibrous dysplasia involved the femoral neck and the intertrochanteric area; in one, it involved the femoral neck only; and in one, it involved the entire proximal part of the femur. The appearance was very similar to that of type-II avascular necrosis, as described by Kalamchi and MacEwen.

Discussion

Lichtenstein[13] is credited with having coined the term fibrous dysplasia in 1938, and, in 1942, he and Jaffe reviewed all known cases of this entity[14]. Those authors established that fibrous dysplasia of bone was a distinct pathological and clinical condition. Since then, there have been many reports about this disease process and its clinical manifestations[10,19,21], particularly McCune-

Albright syndrome[1,15]. However, little is currently known about the pathogenesis of the deformity or the ideal treatment. Studies have shown that a postzygotic somatic mutation of the alpha subunit of the stimulatory G-protein gene, which is responsible for cAMP formation, may cause the abnormal characteristics of tissue in patients who have McCune-Albright syndrome[22]. More recently, Candeliere et al. showed that increased expression of the c-fos proto-oncogene, which is a consequence of increased adenylate cyclase activity, may play a role in the pathogenesis of fibrous dysplasia. The clinical implications of these findings are not yet known.

Fibrous dysplasia has traditionally been divided into three clinical forms: monostotic, polyostotic, and endocrinopathic (McCune-Albright syndrome)[1,15]. It is generally accepted, and we concur, that monostotic lesions are easier to treat, are associated with better outcomes, necessitate fewer operative procedures, and result in fewer microfractures[8]. In the current series, the calcar was not involved in any patient who had a monostotic lesion and the femoral bone was able to support internal fixation in all patients in this group; therefore, no medial displacement osteotomies were performed in the patients who had a monostotic lesion. These patients tended to have less severe deformity, and only one patient had a shepherd's crook deformity. Polyostotic lesions and those associated with an endocrinopathy tend to be larger, with unpredictable results following treatment[8]. In our series, patients who had polyostotic involvement tended to need multiple and more extensive operative procedures, and repeated microfractures were more common. The calcar of the proximal part of the femur often was involved, and more patients had bone that was unable to support internal fixation; thus, medial displacement osteotomies were performed often in this group. A greater degree of varus deformity also was seen in these patients.

Funk and Wells reported that a more predictable, easier course is to be expected in patients who have the monostotic form of the disease. In their study, polyostotic disease involved the proximal part of seven femora in four patients, who had a total of nineteen operations. In patients who had severe polyostotic disease, it was not always possible to control the deformity, even with multiple bone-grafting procedures and osteotomies. For such patients, those authors recommended excision of the involved intertrochanteric area (performed in four femora in their series). They also believed that the best way to correct shepherd's crook deformity was by total excision of the pathological intertrochanteric area, with transposition of the iliopsoas and abductors distally. We disagree with those authors' recommendation that prophylactic bone-grafting be done to prevent deformity, as in our experience such grafts were resorbed.

Curettage and bone-grafting has been the mainstay of treatment for symptomatic and even asymptomatic lesions of fibrous dysplasia. According to our review of the literature, it is uncertain whether this form of treatment offers a definitive solution. Grabias and Campbell believed that curettage or biopsy of an isolated lesion may predispose the bone to pathological fracture or recurrence of the lesion. They also stated that there is no accurate indication of the rate of success of curettage and bone-grafting. In the present series, the fibrous dysplasia was not eradicated in any femur, and no lesion decreased in size following curettage and bone-grafting.

Enneking and Gearen reported their experience in treating symptomatic lesions of the femoral neck with cortical bone-grafting without curettage[6]. They had been dismayed by the results of curettage and autologous cancellous bone-grafting, stating that the grafts were incorporated quickly and eventually were replaced by dysplastic tissue. Nakashima et al. reported that six of their eight patients who had a lesion of the femoral neck were managed successfully with curettage and bone-grafting. However, they did not mention the type of bone that was grafted or the size of the lesions. Those authors concluded that curettage and bone-grafting is the initial procedure of choice for young patients who have a primary lesion in the femoral neck. Harris et al. reported on ten patients who had involvement of the femoral neck; nine were managed with curettage and bone-grafting and one, with curettage and bone-grafting as well as internal fixation. These patients had four good results, one fair result, and five poor results. Strassburger et al. reported that only three of six curettage and bone-grafting procedures resulted in healing of the lesions, which were described as initially being small. DePalma and Dodd reported three successful results after four curettage and bone-grafting procedures in the proximal part of the femur. Stephenson et al. noted better results after curettage and bone-grafting in patients who were eighteen years old or more.

Despite the sporadic good results reported in the literature, the results of the present study call into serious question the role of curettage and bone-grafting in the treatment of these lesions. No lesion in our series was eradicated or diminished in size, and all bone grafts, both cortical and cancellous, were resorbed. Curettage further disrupts what little trabecular support remains in the bone, and it appears that most bone grafts are destined to be resorbed. Enneking and Gearen used cortical autologous grafts from the fibula without curettage of the lesion[6]. A similar operation was performed on one femur with a monostotic lesion in our series (Case 3). Even though whole fibular strut grafts were used, eventually they were resorbed.

DePalma and Dodd reported on six patients who had involvement of the proximal part of the femur. Those authors concluded that medial displacement osteotomy is the procedure of choice for patients who have a shepherd's crook deformity and that valgus osteotomy should be performed in patients who have coxa vara. DePalma and Dodd stated that medial

displacement osteotomy achieves a more functional and serviceable weight-bearing relationship, alleviates stresses and thus helps to prevent pathological fracture, improves stability and gait, and decreases pain. None of their patients had evidence of recurrence or progression of the deformity, either at the site of the original deformity or in an adjacent area. Two patients had coxa vara deformity and were managed with a subtrochanteric osteotomy with medial displacement; both had a satisfactory result[4]. In the series of Harris et al., seven patients who had a shepherd's crook deformity were managed with a subtrochanteric osteotomy, and two also had internal fixation; only two patients had a successful result.

In the present report, osteotomy with internal fixation led to good results. The results were poorer when correction was lost in the postoperative period because of lack of support due to the absence of internal fixation and when the instrumentation did not extend the entire length of the femoral neck. We found that it is important for the neck portion of the internal fixation device to extend into the femoral head in skeletally mature patients and to the level of the physis in skeletally immature patients. No single type of internal fixation appeared to offer a major advantage; equally good results were noted after use of plates, screws, wires, and pins. The surgeon must be flexible with regard to instrumentation as the intraoperative findings often dictate the type of hardware to be used. If internal fixation is not technically feasible, a medial displacement osteotomy, as described by McMurray, may be done. We have found this procedure to be useful, mainly in patients who have involvement of the calcar, with the quality of the bone unable to support internal fixation, and in patients who have a shepherd's crook deformity.

Freeman et al. reported the results of use of the Zickel nail in four patients (six femora) who had polyostotic disease. After one to five years of follow-up, three patients were pain-free. Neither refracture nor progression of the deformity occurred in the five femora that had had adequate fixation. Those authors recommended the Zickel nail because it provides intramedullary stabilization into the distal part of the femur; decreases stress concentration distally; stabilizes the femoral neck with fixation into the femoral head; and, in combination with an osteotomy, can correct deformity. Connolly reported successful results with use of the Zickel nail for the treatment of shepherd's crook deformity. Although the one femur in the present series that was treated with a Zickel nail had a very good result at the 17.6-year follow-up evaluation, we noted thinning of the medial cortex of the shaft, which probably was due to stress-shielding caused by the intramedullary rod. It would be interesting to know whether the long-term results in the series of Freeman et al. included thinning of the medial cortex.

After a pathological fracture, the dysplastic bone has the capability to heal at a rate comparable with that of normal bone in the same anatomical area[5]. However, because the callus consists of fibrodysplastic bone, biomechanical realignment is necessary to provide a valgus position to minimize subsequent fracture and deformity[5]. Of 125 fractures reported on by Harris et al., only two did not unite; those authors believed that most pathological fractures heal well after non-operative treatment and that such treatment should be attempted. Grabias and Campbell stated that the proximal fragment of a fracture should be reduced in a valgus position to provide stable fixation, compensate for any limb-length discrepancy, and prevent recurrent deformity. They also thought that orthopaedic surgeons commonly accept a less-than-optimum position in patients who have extensive involvement; we agree with those authors that this practice should be discontinued.

No patient in our series had a non-union. We believe that most patients who have a fracture of the proximal part of the femur due to fibrous dysplasia can be managed with a one and one-half hip-spica cast provided that a valgus neck-shaft angle is maintained and provided that they are followed diligently with serial roentgenograms to ensure that bowing and varus angulation are not occurring. Curettage and bone-grafting should not be a treatment option for these patients. Patients who have repeated microfractures may need an osteotomy with realignment to place the proximal part of the femur in a more biomechanically favorable valgus position to promote healing. The osteotomy should provide valgus overcorrection (more than 130 degrees) in anticipation of postoperative loss of valgus alignment secondary to bending of the diseased bone. This was not always possible in our patients who had severe polyostotic disease; however, a satisfactory clinical result can be expected when the neck-shaft angle is at least 90 degrees.

The appearance of medial elevation of the epiphysis in four femora in the current series is interesting. Although the roentgenographic appearance is similar to that of type-II avascular necrosis, as described by Kalamchi and MacEwen, we are uncertain whether it is a vascular event or whether it represents extension of the diseased tissue across the lateral aspect of the physis[12]. The data in the literature conflict with regard to whether fibrous dysplasia crosses the physis in skeletally immature patients, whereas its occurrence in skeletally mature patients has been documented[18]. In their large study of the natural history of the disease, Harris et al. found that only two patients had involvement of the capital epiphysis before puberty, although such involvement often occurred after puberty.

No patient in our series had a satisfactory roentgenographic result because no lesion was eradicated or decreased in size. At the time of the latest follow-up, twenty patients (twenty-four femora) did not have a fracture of the femur or incapacitating pain. These patients had a clinically satisfactory result according to our

criteria. However, two patients (three femora) had an unsatisfactory clinical result. Both of these patients had severe polyostotic disease with involvement of the entire proximal part of the femur. One patient, in addition to having bilateral disease, had involvement of almost the entire skeleton and was restricted to a wheelchair. The other patient had a twelve-centimeter limb-length discrepancy and a recurrent shepherd's crook deformity; that patient was able to walk inside the house only, with the aid of crutches.

It is apparent, on the basis of the present series and the literature, that fibrous dysplasia of the proximal part of the femur poses formidable problems for the orthopaedic surgeon. We agree with Jaffe that "the mere fact of their presence is not in itself an indication for treatment [and] only those lesions should be treated which are causing pain or predisposing the bone to fracture." We believe that asymptomatic lesions should not be treated. It is important to remember that the lesions do not cause pain or deformity unless a microfracture develops.

The persistence of repeated microfractures in the pathological bone must be addressed. Our results show that repeated attempts at curettage and bone-grafting do not lead to healing of the lesion and may in fact further weaken the bone because of the manual disruption of the structural trabeculae. Microfractures should be treated with a one and one-half hip-spica cast as the dysplastic bone has the same healing potential as normal bone. However, close observation is needed to detect progression to a varus deformity. Varus deformity should be treated early with a valgus osteotomy and internal fixation; medial displacement should be added if the calcar is involved or if the bone cannot support internal fixation. In skeletally immature patients the internal fixation device in the femoral neck should extend to the growth plate, whereas in adults it should extend into the femoral head. The surgeon must be flexible with regard to the type of internal fixation that is used as the site and location of the lesion and the quality of bone vary among patients. The problems associated with fibrous dysplasia of the proximal part of the femur are not insurmountable if the treatment is carefully planned.

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